Tetralogy of Fallot: Total Correction

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The events that give rise to the development of the constellation of lesions that we refer to as tetralogy of Fallot are still uncertain. Regardless of the true teleological cause, it is helpful for the surgeon to approach this defect as resulting from anterocephalad deviation of the ventricular outlet septum, “monology with sequellae.” The resulting malalignment ventricular septal defect (VSD), aortic override, and right ventricular outflow tract obstruction are thus easily understood. The right ventricular hypertrophy can be appreciated as secondary to the VSD and right ventricular outflow tract obstruction. Right ventricular outflow obstruction is increased by hypertrophy of the anterior limb of the septal band, anomalous anterior muscular bands, and outlet septum as well as pulmonary valve and arterial anomalies (Fig 1).

Classically the VSD is circular, juxtaaortic, and nonrestrictive and results from the malalignment of the parietal extension of the infundibular and trabecular septae. As such, the defect lies between the infundibular septum and is cradled by the anterior and posterior limbs of the septal band (Fig II).

The aorta overrides the VSD, is rotated more clockwise than usual, and is enlarged. Approximately 25% of patients have a right aortic arch, most often with mirror-image branching. The ductus arteriosus is absent in approximately 30% of patients.

The right ventricle is hypertrophied and, as a result of the nonrestrictive VSD, is similar in mass and thickness to the left ventricle. When viewed externally, the right ventricle occupies the entire anterior portion of the heart and the intraventricular septum is displaced to the left. Often the point of maximum narrowing between the inflow and outflow portions of the right ventricle can be seen externally as a transverse indentation on the epicardial surface during ventricular systole.

Right ventricular outflow tract obstruction can occur at multiple levels within the ventricle (Fig III). Right ventricular outflow tract obstruction is further complicated by variable hypoplasia of the pulmonary valve annulus and pulmonary arteries themselves. Valvar pulmonary stenosis occurs in approximately 75% of the patients, often in concert with hypoplasia of the pul-

Fig I. Anatomy of tetralogy of Fallot. Displacement of the infundibular septum (IS), parietal extension (PE), and septal extension (SE) results in a malalignment VSD, right ventricular outflow tract obstruction (RVOTO), and right ventricular hypertrophy.
**Fig 11.** (A) Normally, the conal septum (infundibular septum [IS] inserts between the anterior and posterior limbs of the septal band (SB). (B) In tetralogy of Fallot the conal septum is deviated both anterior and superior, resulting in a malalignment VSD and right ventricular outflow tract obstruction. (Adapted with permission.)

**Fig III.** Anatomic variations in tetralogy of Fallot: (A) localized midinfundibular obstruction with distal infundibular chamber (IC); hypertrophic long (B) and short (C) infundibular muscle obstruction; hypoplastic long (D) and short (E) infundibular obstruction.
monary valve annulus and supravalvar obstruction at the level of the sinotubular ridge.

The left ventricle is usually of normal thickness. The coronary arteries become quite dilated and tortuous over time. Often a large conal branch of the right coronary artery runs across the free wall of the right ventricle. Significant coronary artery anomalies have been reported in approximately 5% of patients with tetralogy of Fallot. Most commonly, the left anterior descending artery arises from the right coronary artery and crosses the right ventricular outflow tract. This anomaly must be recognized when planning relief of right ventricular outflow tract obstruction, because it may limit the ability to perform an effective transannular incision, possibly necessitating the use of a right ventricle-to-pulmonary artery conduit.

The vast majority of patients with tetralogy of Fallot have a nonrestrictive VSD, producing systemic RV pressures. The net shunt at the ventricular level is then dependent on the degree of right ventricular outflow tract obstruction.

Approximately 70% of patients with tetralogy of Fallot and pulmonary stenosis require surgical intervention in infancy because of hypoxic spells or persistent and progressive systemic hypoxemia. Debate continues on the appropriate use of initial palliation as opposed to primary repair, as well as the optimal timing for a complete repair in the asymptomatic infant. Currently, most major centers are opting for complete repair in the absence of some relative contraindication. These may include multiple VSDs, associated atrioventricular septal defect, important coronary artery anomalies, very small branch pulmonary arteries, and extreme tubular infundibular hypoplasia requiring a long ventriculotomy and transannular patch to correct. In these cases a standard Blalock-Taussig (BT) shunt can provide excellent palliation and allow for interval repair. Rarely, emergent operation may be required in neonates with severe cyanosis and the inability to maintain a patent ductus arteriosus. In an extremely desaturated patient, a central aortopulmonary shunt can be performed while the patient is on cardiopulmonary bypass, allowing for correction of hypoxia, acidosis, and other metabolic derangements.
The heart is approached through a median sternotomy. The thymic tissue is excised as necessary to provide an unobscured view. The pericardium is opened in the midline, and a pericardial tent is created. A decision is then made concerning cannulation techniques. In neonates and small infants, the surgeon may prefer single atrial venous cannulation and the use of profound hypothermia, low-flow bypass, and a period of circulatory arrest to optimize visualization of the intracardiac structures. The remainder of the patients are treated with standard bivacal venous cannulation and moderate hypothermia (28-30°C). The use of a left heart vent, placed either through the right superior pulmonary vein or across the atrial septum, can be helpful in maintaining a bloodless field during resection of the infundibular obstruction and closure of the VSD.

Once cardiopulmonary bypass is established, mobilization of the proximal pulmonary arteries (including division of the patent ductus or ligamentum arteriosum) can be helpful in addressing branch pulmonary artery stenosis and pulmonary valvar/annular hypoplasia. (A) After aortic cross-clamping, a dose of cold blood cardioplegia (1:1, 15 ml/kg) is administrated and repeated if the cross-clamping time exceeds 20-30 minutes. An oblique right atriotomy is fashioned from a point posterior to the right atrial appendage paralleling the atrioventricular groove and remaining anterior to the IVC orifice. The anterior and septal leaflets of the tricuspid valve are gently retracted toward the surgeon. Using a curved blunt instrument, the right ventricular outflow tract is probed to confirm the proper pathway from the right ventricular cavity to the main pulmonary artery. (B) In neonates and infants, simple division of obstructing muscle bundles is carried out from the level of the moderator band to the pulmonary valve annulus along the intraventricular septum. As the division of these muscle proceeds, the right ventricular free wall is freed from anomalous attachments to the intraventricular septum, providing a better view of the distal right ventricular infundibulum. The parietal extension of the infundibular septum can then be visualized and pulled into the operative field, with a portion of muscle excised if necessary. Care must be taken to avoid injury to the aortic valve leaflets and the margin of the VSD. Any obstructive fibrous tissue in the outflow tract is also excised; however, the surgeon should avoid cutting muscle bundles that are not obstructive, especially in the body of the right ventricle, to preserve right ventricular function. After these maneuvers, the inferior aspect of the pulmonary valve leaflets should be clearly visualized. Hegar dilators are then passed through the tricuspid valve, out the right ventricular infundibulum, and across the pulmonary valve annulus, to calibrate and confirm enlargement of the right ventricular outflow tract.
After division of the tethering right ventricular muscle bundles, the VSD margins are now more easily seen. The defect is closed using a patch (polytetrafluoroethylene [PTFE]), usually circular in shape, and approximating the size of the aortic valve annulus. A continuous running suture technique is used, beginning at the point farthest from the surgeon's view. This corresponds to the transected end of the parietal extension of the infundibular septum. Sutures are placed through this muscle and continue over and across the anterior aspect of the aortic valve annulus. Here they are placed in close proximity to the aortic valve leaflets, to prevent residual shunting through right ventricular trabeculations. This arm of the suture continues to the crux of the heart, at which point it passes from the ventricular to the atrial side through the base of the tricuspid valve near its anteroseptal commissure. Aortic valve competency can be evaluated by giving a "test dose" of cardioplegia while directly inspecting the valve leaflets. Around the posteroinferior margin, the VSD sutures are placed approximately 2 mm from the edge, to avoid injuring the conduction tissue. The suture then transitions from the muscular septum through the base of the septal leaflet of tricuspid valve. With a horizontal mattress technique, the patch is attached to the base of the tricuspid valve septal leaflet until the first suture placed through the anteroseptal commissure is met. The sutures are tied over a pericardial pledget to prevent tearing through the septal leaflet tissue. The right ventricular cavity is instilled with cold saline, and the competency of the tricuspid valve is tested. Occasionally, a commissuroplasty in the anteroseptal commissure can help alleviate any residual tricuspid insufficiency. Any significant atrial defect is enclosed after the left atrium is deaired.
The surgeon's attention is then directed to the main and branch pulmonary arteries and the distal right ventricular outflow tract. The main pulmonary artery is opened longitudinally, and the branch pulmonary arteries are inspected. The pulmonary valve is examined; it is often bicuspid, thickened, and fused at the commissures. A pulmonary valvotomy is performed, the pulmonary valve annulus is sized, and the right ventricular infundibulum is visualized through the pulmonary valve. Any residual distal right ventricular outflow obstruction (ie, distal parietal insertion of the infundibular septum) is resected through the pulmonary valve annulus. At this point in the repair, the surgeon must decide whether the right ventricular outflow tract size (volume) will be sufficient to allow for the entire cardiac output to traverse it without causing extreme elevation in right ventricular pressure. The pulmonary valve annulus size when related to the child’s size is expressed as normalized “Z values.” In short, the Z value represents the number of standard deviations above (+) or below (-) the mean normal values for age and size.

In general, a transannular patch can be avoided if the pulmonary valve annulus is larger than a Z value of -2. Patients with more proximal uncorrectable right ventricular obstruction (ie, diffuse tubular hypoplasia of the infundibulum, in concert with pulmonary annular hypoplasia) may require a subannular or transannular patch to effectively relieve the length of this obstruction. If the obstruction is confined solely to the pulmonary valve annulus, a very limited (3-4 mm) incision onto the right ventricular outflow tract may be sufficient.

Fashioning the transannular patch correctly is important to relieve the obstruction and prevent excessive regurgitation. The patch should be oval-shaped with blunt corners and may be composed of autologous or preserved pericardium as well as PTFE. A Hegar dilator, 2-3 mm larger than the mean normal-sized pulmonary annulus, is used to appropriately size the patch. The maximal width of the patch should be slightly larger than the portion of the circumference of the Hegar dilator visible in the open right ventricular outflow tract.

In the presence of an anomalous left anterior descending coronary artery crossing the right ventricular outflow tract, the transatrial/transpulmonary approach is still used. The position and course of the coronary will limit the ventriculotomy that can be performed. A short (2-4 mm) transannular patch often can be used and may be sufficient. We do not recommend mobilizing the vessel and subcoronary patch but will accept slightly higher RV pressures in this situation. If aggressive transatrial muscle division/resection does not adequately address the obstruction, then an right ventricle-pulmonary artery conduit may be necessary. If this information is known preoperatively, some surgeons will prefer to defer complete repair in the neonate or infant. A BT shunt can be performed, followed by a complete repair in the hope of placing a larger conduit. The cross-clamp can be removed during patch insertion and the patient rewarmed. Finally, the right atriotomy is closed.
Comments

Survival after repair of tetralogy of Fallot is excellent; however, common postoperative problems include hemodynamic derangements and arrhythmias. In general, patients with cyanotic congenital heart defects are thought to have abnormal systemic and pulmonary capillary membranes, which may make them more susceptible to changes incurred during cardiopulmonary bypass. Patients with tetralogy of Fallot are not only cyanotic but may have intrinsic myocardial and pulmonary abnormalities that can further complicate their care.

Forty years after the first successful repair of tetralogy of Fallot, surgical techniques continue to evolve and controversies persist. Many centers are reporting excellent results with complete repair in the first few months of life. Earlier repair may avoid the effects of chronic cyanosis, prevent the progression of right ventricular hypertrophy and fibrosis, and encourage development of the pulmonary bed.

REFERENCES


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